

## Expected Practices

Specialty: Rheumatology

Subject: Approach to Scleroderma

Date: May 20, 2014

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### **Purpose:**

Approach to the diagnosis and initial management of Scleroderma

**Target Audience:** Primary Care Physicians

### **Expected Practice:**

#### *When to think of Scleroderma*

The scleroderma spectrum of diseases includes diffuse cutaneous scleroderma and limited cutaneous scleroderma. These diseases result from fibroblast dysfunction causing increased deposition of extracellular matrix into the skin and other organs. The result is skin thickening and often internal organ involvement. Signs and symptoms of scleroderma may include thickening of the skin of the fingers and hands, severe Raynaud's, gastroesophageal reflux, dyspnea due to interstitial lung disease or pulmonary hypertension, diarrhea, arthritis, telangiectasias on the skin, and occasionally subcutaneous deposits of calcium. Diffuse cutaneous scleroderma presents with progressive skin thickening starting in the fingers and hands and progresses to include skin of the trunk. These patients are at higher risk for interstitial lung disease and renal involvement. Limited cutaneous scleroderma usually presents with longstanding severe Raynaud's followed by skin thickening later in the course. The skin thickening is limited to below the elbows, knees, and above the clavicles, not involving the trunk. Consider scleroderma in any patient that presents with these features.

#### *How to test and risk stratify for Scleroderma and when to refer*

Start your evaluation with a history focusing on the duration of symptoms. Inquire about a family history of autoimmune diseases or scleroderma. Ask about skin thickening of the fingers or face, Raynaud's, shortness of breath, and gastroesophageal reflux. On physical exam check the fingers, hands, and face for

*This Expected Practice was developed by a DHS Specialty-Primary Care Work Group to fulfill the DHS mission to ensure access to high-quality, patient-centered, and cost-effective health care. SPC Work Groups, composed of specialist and primary care provider representatives from across LA County DHS, are guided by 1) real-life practice conditions at our facilities, 2) available clinical evidence, and 3) the principle that we must provide equitable care for the entire population that LA County DHS is responsible for, not just those that appear in front of us. It is recognized that in individual situations a provider's clinical judgment may vary from this Expected Practice, but in such cases compelling documentation for the exception should be provided in the medical record.*

skin thickening (see below) and telangiectasias. When examining the hands, look for Raynaud's, swollen and tender joints, ischemic pitting ulcers on the fingertips (see below), and evaluate the nail folds for evidence of abnormal capillary dilation or dropout (see below). Listen to the heart and lungs for the stigmata of pulmonary hypertension or interstitial lung disease.



The diagnosis of scleroderma is largely clinical with very little reliance on serologic tests (see reference below). Anti-Scl-70, anti-centromere, and anti-RNA polymerase III can be helpful, but are poorly sensitive and thus not required prior to referral to rheumatology.

Referral to rheumatology should be made in all patients with a characteristic history and examination findings as discussed previously.

**\*For those patients with acute or subacute onset of severe shortness of breath or new onset malignant hypertension; prompt referral to the emergency room is indicated for expedited diagnosis and management.**

#### *Initial management of newly diagnosed Scleroderma*

eConsult to Rheumatology is recommended for patients with newly diagnosed scleroderma. If managing a patient with organ specific manifestations, the following information should be reviewed.

1. Pulmonary fibrosis: Acquire a chest x-ray and refer to rheumatology for further evaluation and consideration of immunosuppressive therapy.
2. Pulmonary Artery Hypertension: screen with transesophageal echocardiography and consider a right heart catheterization if indicated to establish the diagnosis. Recommend expedited referral to rheumatology to coordinate management with pulmonary and/or cardiology services.
3. Renal: Patients with scleroderma are at risk of developing scleroderma renal crisis. Prompt hospitalization and management with ACE-inhibitors is indicated if patient develops abrupt elevation in blood pressure, renal failure, proteinuria/microscopic hematuria, or microangiopathic hemolytic anemia. Avoid using doses of prednisone > 15 mg daily as this has been associated with an increased risk of developing scleroderma renal crisis. Do not use ACE-inhibitors prophylactically in all scleroderma patients as this has been associated with increased mortality. Have patients monitor blood pressures daily with a home blood pressure cuff.
4. Raynaud's syndrome: Hand warming measures/gloves, keep core temperature warm, and consider using long acting dihydropyridine calcium channel blockers.
5. Gastroesophageal Reflux Disease: Treat with H<sub>2</sub> blockers or proton pump inhibitors.